

Renal hemosiderosis complicating sickle cell anemia

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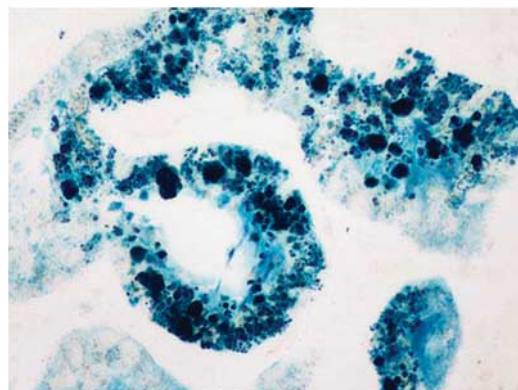


Figure 1 | Perls staining revealing intense hemosiderosis involving the renal tubules.

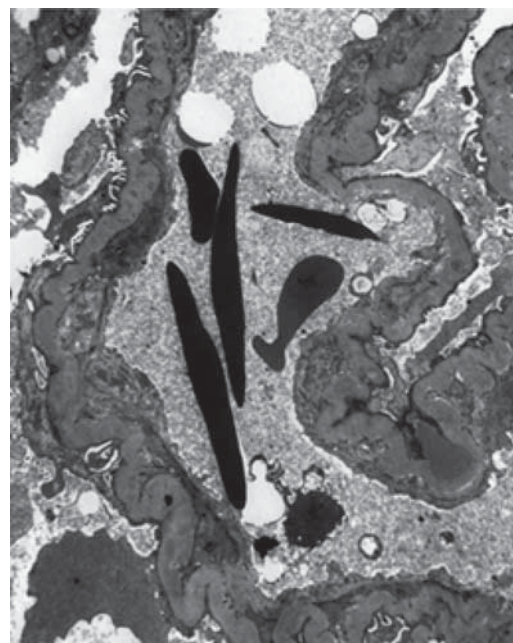


Figure 2 | Electron microscopy showing sickle red cells inside a glomerular capillary loop.

A 37-year-old Brazilian black male with a history of sickle cell anemia presented with a 2-week history of pain and edema in the lower extremities, accompanied by generalized weakness and dyspnea on exertion. Outpatient laboratory evaluation revealed hemoglobin of 4.3 mg/dl, creatinine of 5.8 mg/dl, and blood urea nitrogen of 100 mg/dl. Ten months prior to admission, he had been hospitalized for a hypertensive crisis; at that time, his serum creatinine was 1.3 mg/dl. Upon admission to our service, he was pale and slightly jaundiced; blood pressure was 180 × 100 mm Hg. A workup for the renal dysfunction revealed 1+ protein on dipstick and 30 red cells per high-power field on urine sediment; renal ultrasound showed normal-sized kidneys but with increased echogenicity; viral serologies (human immunodeficiency virus, hepatitis C virus, hepatitis B virus) and lupus studies

(antinuclear antibody, anti-DNA, anti-SM) were negative. A renal biopsy revealed intense hemosiderin deposits in the renal tubules (Figure 1) and sickle red cells in glomerular capillaries (Figure 2). Iron studies, liver panel, and serum lipids were within normal limits; hemoglobin electrophoresis confirmed SS hemoglobinopathy. Hemolysis studies were not performed. The patient progressed to end-stage renal disease and is currently on maintenance hemodialysis. Renal hemosiderosis is a rare cause of renal failure that can occur in diseases characterized by chronic intravascular hemolysis, such as paroxysmal nocturnal hemoglobinuria. In sickle cell anemia, abnormal red cells are usually destroyed in the spleen (extravascular) but intravascular hemolysis may occur during acute crises. Free hemoglobin is filtered by the glomeruli and reabsorbed by proximal convoluted tubules, leading to renal hemosiderosis.